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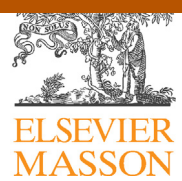

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CLINICAL RESEARCH

Repeat mitral valve repair for haemolysis in children

Chirurgie redux pour hémolyse après plastie chirurgicale de la valve mitrale de l'enfant

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KEYWORDS
Reoperation;
CHD valve;
Mitral valve repair

Summary
Background. — Severe haemolysis is a rare complication after mitral valve repair in congenital heart disease.
Aim. — We describe four children with severe mitral regurgitation who underwent valve repair and subsequently developed profound haemolytic anaemia.
Methods. — Clinical, echocardiographic and surgical data were collected retrospectively from a surgical centre in France during a 5-year period.
Results. — Two patients had atrioventricular septal defects, one patient had congenital mitral dysplasia and one had anomalous left coronary artery from the pulmonary artery with mitral regurgitation. Haemolysis was diagnosed 20 to 75 days after surgery, as a result of clinical and biological examination; it was severe, and blood transfusion support was necessary in all cases. Haemolysis was always associated with eccentric mitral regurgitation with a variable degree of severity (from low to severe). After exclusion of other haemolysis aetiology, redo mitral repair surgery was performed successfully in all cases. The haemolysis was considered to be mechanical in origin, caused by regurgitation of blood through the residual mitral regurgitation and stitches.

Abbreviations: AVSD, Atrioventricular septal defect; LAVV, Left atrioventricular valve; MR, Mitral regurgitation.

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Conclusion. — Severe haemolysis is a rare complication that can occur after mitral valve repair in congenital heart disease. All patients underwent successful redo repair and had resolution of haemolysis after surgery. Even if redo surgery is required, iterative mitral repair is possible.

MOTS CLÉS
Réopération ;
Cardiopathie congénitale ;
Valvuloplastie

Résumé
Contexte. — La survenue d'une hémolyse après chirurgie mitrale est une complication rare et décrite le plus souvent après remplacement valvulaire mécanique.
Objectif. — Cette étude décrit la survenue de quatre cas d'anémies hémolytiques symptomatiques survenues après plastie mitrale chirurgicale chez des enfants opérés pour diverses cardiopathies congénitales.
Méthodes. — Les données cliniques, biologiques et chirurgicales ont été collectées de façon rétrospective dans le centre de chirurgie cardiaque pédiatrique Marie-Lannelongue sur une période de cinq ans.
Résultats. — Les enfants avaient un canal atrio-ventriculaire dans deux cas, une dysplasie mitrale isolée dans un cas et une naissance anormale de la coronaire gauche à partir de l'artère pulmonaire associé à une insuffisance mitrale dans le dernier cas. Tous ces enfants ont développé une anémie hémolytique symptomatique. Le diagnostic de cette complication a été clinique et biologique. Tous les enfants avaient une insuffisance mitrale résiduelle de grade léger à sévère. Tous les enfants ont nécessité des transfusions sanguines. Après exclusion de toute autre cause, tous les enfants ont bénéficié d'une nouvelle plastie mitrale chirurgicale. Cette nouvelle intervention a permis la correction de l'hémolyse dans tous les cas. Il s'agissait d'hémolyse mécanique liée à une fuite mitrale résiduelle ou une fuite entre les sutures.
Conclusion. — La survenue d'une anémie hémolytique est une complication rare de la plastie mitrale chirurgicale chez l'enfant. La reprise chirurgicale permet de traiter l'hémolyse. Quand une ré-intervention est nécessaire, une nouvelle plastie mitrale reste possible.

Background

Congenital mitral valve disease presents with a wide variety of morphological abnormalities and a high incidence of associated intracardiac anomalies. In light of improvements in surgical techniques, repair rather than replacement is preferred because of the deleterious effects associated with prosthetic valve placement in small children [1]. While haemolysis after mitral repair (and its remedial management) has been reported in adults [2,3], it has been described rarely for isolated mitral disease in children [4].

Methods

Since 2006, 493 mitral repair procedures in children aged < 15 years were performed at our institution (100 partial, 249 complete atrioventricular septal defects [AVSDs], 144 isolated congenital mitral valve diseases). We detail our experience and management strategy in four children with post-repair haemolysis. No other case of severe haemolysis was observed in the centre's experience.

In all four children, blood tests demonstrated haemolytic anaemia without thrombotic microangiopathy; they all underwent blood transfusions and received beta-blockers (except patient 4), but the condition did not improve. Intraoperative trans-oesophageal echocardiography was used systematically in the repair procedures.

Results

Case descriptions

Patient 1

A newborn was diagnosed with a partial AVSD, right chamber dominance and aortic arch obstruction. After neonatal arch repair (Crafoord through thoracotomy), the atrial septal defect was closed at day 14, because of large symptomatic shunt, without left cleft closure, as no regurgitation was found at the time of surgery. The child progressively developed congestive heart failure due to severe left atrioventricular valve (LAVV) regurgitation and subaortic stenosis. Cleft suture and subaortic enlargement were performed at 8 months. Twenty-two days later, the patient was admitted for macroscopic haematuria. Echocardiography demonstrated two new-onset LAVV jets, one directed towards the septum and one towards the lateral atrial wall (Fig. 1). Three weeks after admission, completion of LAVV cleft closure was performed with subsequent resolution of haemolysis.

Patient 2

A 2-year-old girl presented with a partial AVSD with significant right chamber dilatation. Closures of the LAVV cleft and the ostium primum were performed. Postoperative echocardiography showed mild regurgitation. Two months later, the

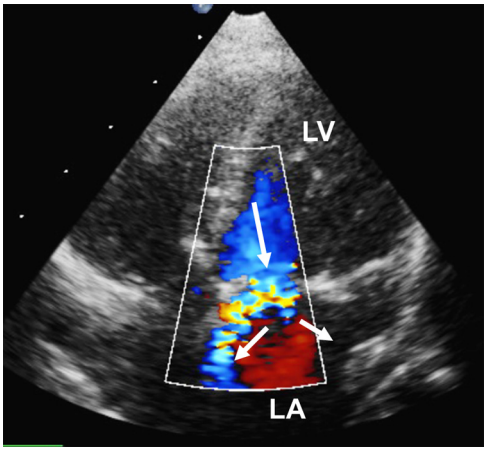

Figure 1. Transthoracic apical view showing two regurgitation jets (arrows) (rapid acceleration and fragmentation mechanisms) on a residual cleft (patient 1). LA: left atrium; LV: left ventricle.

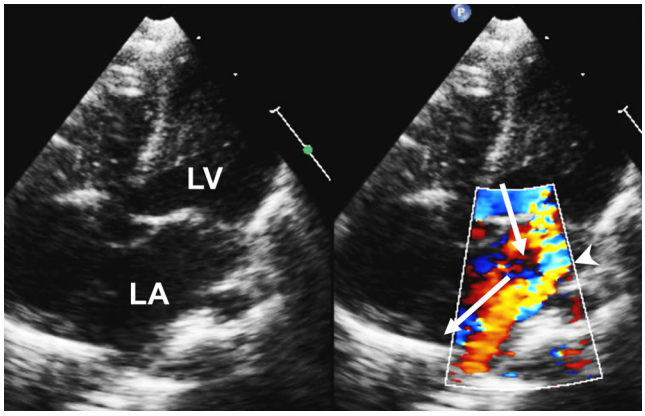

Figure 2. Transthoracic apical view showing severe regurgitant jet (arrows) colliding (arrowhead) with the posterior atrial wall (patient 2). LA: left atrium; LV: left ventricle.

patient was admitted with macroscopic haematuria. Regurgitation was severe, with a jet towards the lateral atrial wall due to lack of co-aptation (residual cleft) (Fig. 2). LAVV repair was performed 4 months later and included removal of two sutures suspected to be responsible for haemolysis, annuloplasty, commissuroplasty and subvalvular debridement. Haemolysis resolved postoperatively.

Patient 3

A 3-month-old girl was admitted with congestive heart failure. Anomalous left coronary artery arising from the pulmonary artery was diagnosed, associated with severe ischaemic mitral regurgitation (MR). Retraction of the posterior leaflet, annular distortion and brightness of the anterolateral papillary muscle were observed. Surgery was performed, with coronary relocation in the aorta and mitral repair (posterior annuloplasty). Twenty days later, the patient was admitted with haematuria; mild central regurgitation was found. Redo surgery was performed 5 weeks later, with subvalvular debridement and anterior leaflet patch augmentation, which eliminated MR and led to a resolution of haemolysis.

Patient 4

A 3-month-old boy was admitted for heart failure. Aortic arch obstruction and mitral dysplasia were diagnosed. Severe MR was caused by a co-aptation defect, with restrained posterior leaflet by abnormal basal chordae. Mitral (posterior leaflet debridement) and aortic arch repair were performed. Two weeks later, the patient was admitted with symptomatic haematuria. Echocardiography showed moderate MR, with a jet divided by a ruptured chordae on the posterior leaflet. Redo surgery was performed within a month and consisted of anterior leaflet patch augmentation, which eliminated MR and led to the subsequent resolution of haemolysis.

Discussion

With the improvement of surgical materials and techniques, clinically significant haemolysis, caused by intravascular or intracardiac prosthesis, has become uncommon. In congenital heart surgery, the majority of reported cases have involved foreign materials. In the absence of prosthetic material, five cases were described: one caused by coarctation, which resolved after repair [5], one resulting from pulmonary branch stenosis after an arterial switch operation [6] and three occurring after AVSD repair [7—9]. In these cases, no prosthetic material was used and the sources of haemolysis were the residual defects, resulting in regurgitation. Using echocardiography, Yeo et al. [10] described the mechanisms of haemolysis after repair: fragmentation (jet divided by a solid structure, suture or dehiscent ring), collision (direct impact on a solid suture), rapid acceleration (through a small orifice [leaflet perforation, ring dehiscence]), free jet (through a wide orifice) and slow deceleration (through an eccentric orifice, along the atrial wall). Patients 1 and 2 presented rapid acceleration, with a narrow hypervelocimetric jet on a small residual cleft, while patient 3 exhibited the free jet mechanism. Collision was found for patient 2, on the posterior atrial wall, and fragmentation for patient 4, on a ruptured chordae.

Haemolytic anaemia is a rare condition after mitral repair in children, and its occurrence should prompt investigations for recurrent regurgitation. It should be emphasized that all causes of haemolysis, especially in the paediatric field, must be ruled out before a mechanical cause can be confirmed.

In the setting of mechanical haemolysis, anaemia increases cardiac output, and thus, turbulent blood flow and shear stress, and further aggravates intravascular haemolysis. Some reports used this tachycardia-mediated increase to support the use of beta-blockers [11]. Despite beta-blockers being administered in three of our patients, all subsequently required redo surgery.

Of the three haemolysis cases after AVSD repair reported in the literature, two eventually required valve replacement [7,8], while a 70% replacement rate is reported for adult post-repair haemolysis. This percentage varies between series from 0% to 100%, depending on the centre and the type of patients. In our experience, a repeat repair was always possible and cured the haemolysis, provided that regurgitation was eliminated. Intraoperative echocardiography

is thus mandatory to validate the success of the repair. Despite successful reports in the paediatric field, mitral replacement is still associated with persistent problems (restrained annular growth, haemolysis, perivalvular leak, prosthetic thrombosis, thromboembolism and lifelong cumulative risk of anticoagulation) and technical difficulties (extra-anatomic positioning). We recently reported our experience with repair of MR in children (106 patients from 2001 to 2012, during a follow-up period of 3.9 ± 3.2 years [range, 2 months to 11 years]); the reoperation rate in this series was 23% [12]. A clear repair-oriented policy [1] in redo valve surgery is of crucial importance, and, most of time is possible. In these patients, valvular material is stiffened by iterative procedures, and specific repair of residual lesion is often not sufficient. The key procedure is restoration of leaflet mobility. We commonly use extensive subvalvular debridement (secondary chordae cutting) and leaflet patch augmentation, using autologous pericardium (untreated, as the material is already thickened by redo adhesions).

Conclusion

In summary, this report suggests that redo mitral repair is feasible in children with haemolysis related to residual regurgitation. Despite redo, mitral repair is a challenging procedure; surgery cured haemolysis in all children without need for replacement. Haemolysis after mitral repair for congenital disease was thus not associated with worsening prognosis. A diagnostic tool to identify residual lesions at risk needs to be developed.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References

[1] Oppido G, Davies B, McMullan DM, et al. Surgical treatment of congenital mitral valve disease: midterm results of a repair-oriented policy. J Thorac Cardiovasc Surg 2008;135:1313—20 [discussion 20—1].
[2] Lam BK, Cosgrove DM, Bhudia SK, Gillinov AM. Hemolysis after mitral valve repair: mechanisms and treatment. Ann Thorac Surg 2004;77:191—5.
[3] Noble S, Jolicoeur EM, Basmadjian A, et al. Percutaneous paravalvular leak reduction: procedural and long-term clinical outcomes. Can J Cardiol 2013;29:1422—8.
[4] Gupta U, Valencia G, Khan MS, Morales M. A rare case of hemolytic anemia in a pediatric patient due to ring dehiscence after mitral valve repair: utility of real-time three-dimensional imaging and management. Pediatr Cardiol 2014;35:180—2.
[5] Ravenel SD, Johnson JD, Sigler AT. Intravascular hemolysis associated with coarctation of the aorta. J Pediatr 1969;75:67—73.
[6] Schroeder J, Albert J, Clarke D, Schaffer M, Wolfe R, Hays T. Hemolysis due to branch pulmonary stenosis after the arterial switch procedure. Ann Thorac Surg 1991;51:491—2.
[7] Capouya ER, Laks H, Drinkwater Jr DC, Pearl JM, Milgalter E. Management of the left atrioventricular valve in the repair of complete atrioventricular septal defects. J Thorac Cardiovasc Surg 1992;104:196—201 [discussion—3].
[8] Moisey CU, Manohitharajah SM, Tovey LA, Deverall PB. Hemolytic anemia in a child in assoication with congenital mitral valve disease. J Thorac Cardiovasc Surg 1972;63:765—8.
[9] Tsang JC, Shum-Tim D, Tchervenkov CI, Jutras L, Sinclair B. Hemolytic anemia after atrioventricular septal defect repair without synthetic material. Ann Thorac Surg 1999;68:1838—40.
[10] Yeo TC, Freeman WK, Schaff HV, Orszulak TA. Mechanisms of hemolysis after mitral valve repair: assessment by serial echocardiography. J Am Coll Cardiol 1998;32:717—23.
[11] Okita Y, Miki S, Kusuhara K, Ueda Y, Tahata T, Yamanaka K. Propranolol for intractable hemolysis after open heart operation. Ann Thorac Surg 1991;52:1158—60.
[12] Kalfa D, Vergnat M, Ly M, et al. A standardized repair-oriented strategy for mitral insufficiency in infants and children: Midterm functional outcomes and predictors of adverse events. J Thorac Cardiovasc Surg 2014;148:1459—66.